Single Lung Transplantation With Concomitant Cardiac Surgery in a Patient With Cystic Fibrosis: A Case Report

Remigiusz Antończyk[a,b], Maciej Urlik[a], Magdalena Latos[a,b,*], Anastazja Pandel[a,b], Martyna Gawęda[a,b], Mirosław Nęcki[a], Michał Kręt[a], Piotr Przybyłowski[a,c], Marian Zembala[a,b], Marek Ochman[a,b], and Tomasz Stącel[a]

[a]Silesian Center for Heart Diseases, Zabrze, Poland; [b]Department of Cardiac, Vascular, and Endovascular Surgery and Transplantology, Medical University of Silesia, Katowice, Poland; and [c]First Chair of General Surgery, Jagiellonian University, Medical College, Kraków, Poland

ABSTRACT

Background. Cystic fibrosis is a congenital, progressive disease affecting many organs. It frequently leads to severe respiratory failure, which can be treated by means of a double lung transplantation. Single lung transplantation is justified only in certain cases.

Case report. This is a case report describing a 20-year-old female patient who became the recipient of a single lung transplant as a result of cystic fibrosis. The transplant was performed during cardiothoracic surgery, which included an intervention in the right atrium. At the age of 14, the patient underwent left pneumonectomy. In addition, the patient had a percutaneous endoscopic gastrostomy placed and a vascular port implanted. During preoperative evaluation, she presented with clinical symptoms of chronic respiratory failure. The patient was approved for lung transplantation at the age of 16. After 2 years on the national lung transplant waiting list, in 2018, the patient underwent right lung transplantation and removal of numerous thrombi in the right atrium during 1 procedure. This surgery was accomplished with the use of extracorporeal circulation, which is an extracorporeal membrane oxygenator combined with cardiopulmonary bypass. The patient was discharged 3 weeks after the procedure in good general condition. Presently, her pulmonary function is excellent and she presents with normal respiratory capacity.

Conclusions. Patients with cystic fibrosis often require double lung transplantation. Under normal circumstances, performing only a single lung transplantation would be considered medical malpractice. However, in certain cases, a single lung transplant is a life-saving procedure.

CYSTIC fibrosis (CF) frequently leads to severe respiratory failure, which can be treated by means of lung transplantation (LT). Double lung transplantation (DLT) remains the gold standard for patients with CF [1,2]. Such treatment increases life expectancy and improves pulmonary function as measured by spirometry and the 6-minute walk test [3]. Single lung transplantation (SLT) is justified only in certain cases, such as a history of pneumonectomy years before possible LT. Because the only lung must be replaced during the procedure, proper oxygenation is maintained by devices like cardiopulmonary bypass (CPB) or extracorporeal membrane oxygenation (ECMO). If such support is needed, ECMO has been proved to be superior to standard CPB [4]. However, the following case report depicts an unusual patient case requiring resourcefulness and skill in planning the procedure.

CASE PRESENTATION

This case report describes a 20-year-old patient who is the recipient of an SLT as a result of end-stage pulmonary failure in course of CF. Her medical history before and after the procedure is presented in Fig 1. The patient was born in 1999 and was diagnosed with CF at

*Address correspondence to Magdalena Latos, MD, Medical University of Silesia, Silesian Center for Heart Diseases, Ulica Marii Skłodowskiej-Curie 9 Pawilon C, 41-800 Zabrze, Poland. Tel: +48667288893. E-mail: latos.magdalena93@gmail.com

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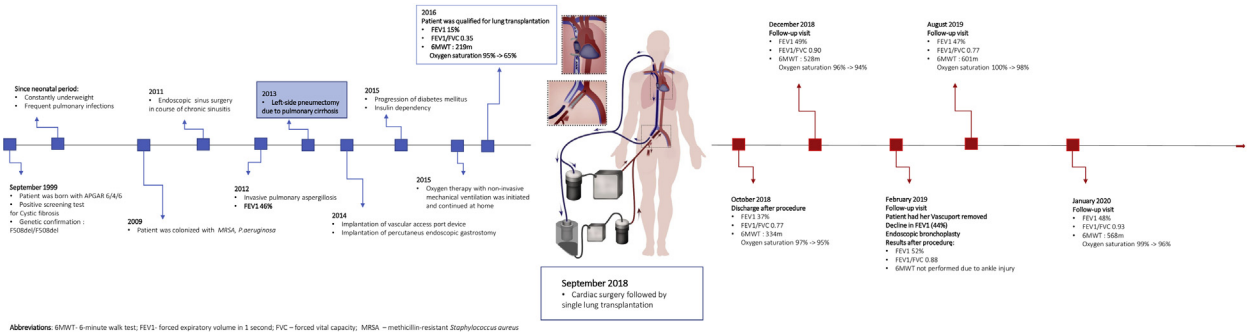

Fig 1. Medical history of a patient with distinction between the pretransplant and post-transplant period.

the age of 6 weeks, as it was revealed that she is a carrier of a double F508del mutation on chromosome 7. During the course of her underlying disease, the patient suffered from recurrent infections of the respiratory tract. Up to the time of her LT, she was always malnourished and underweight. At the age of 10, she became colonized with *Pseudomonas aeruginosa* and methicillin-resistant *Staphylococcus aureus*. At the age of 13, the patient was treated with intravenous voriconazole due to invasive respiratory aspergillosis (*Aspergillus fumigatus*), which eventually caused significant pulmonary cirrhosis followed by left-side pneumonectomy. Histopathological examination of the removed lung confirmed the irreversible pulmonary cirrhosis. Her state was gradually deteriorating. She required implantation of a vascular port and percutaneous endoscopic gastrostomy in her early teens. As it was mentioned previously, owing to malnutrition and fat-soluble vitamin deficiency (mainly vitamin D), she developed scoliosis, as depicted on Fig 2. Her medical history was also significant for diabetes mellitus and nephrolithiasis. Before the patient was referred to the lung transplant facility, her pulmonary disease was progressing and included episodes of hemoptysis and frequent hospitalizations with intravenous administration of antibiotics. The patient was referred for LT approval at the age of 16. At the time of her preoperative evaluation, she was oxygen dependent and presented with 15% of predicted forced expiratory volume in 1 second with severe pulmonary obstruction with a 35% Tiffeneau-Pinelli index. During the 6-minute walk test, she experienced a significant decrease in oxygen saturation from 92% to 65% and reached a distance of only 219 m. Her psychological assessment presented her as a compliant candidate for such a procedure. At the age of 19, a matching donor was reported and the patient was admitted to undergo single LT. At the time of admission for LT, the patient underwent routine echocardiographic assessment. This examination revealed indirect features of pulmonary hypertension, including right ventricular enlargement with a systolic pressure of 44 mm Hg, hypokinesis of the free wall, and mild tricuspid valve regurgitation. Additionally, a polymorphic tumor in the right atrium was suspected. Further radiologic diagnostics were performed including a chest computed tomography, which confirmed the presence of a tumor (Fig 3A). This examination presented left-sided displacement of the mediastinum, remnants of the left pulmonary artery and pulmonary veins, and properly developed pulmonary circulation on the other side (Fig 3B). Her remaining lung had substantial bronchiectasis and was filled with dense mucus. It was decided that cardiac surgery will be followed by SLT. Such a procedure required a resourceful approach to cardiopulmonary support, and 3 stages of the procedure are depicted in Fig 4 (please pay attention to the placement of the clamps while reading about the stages of the procedure). Combined ECMO and CPB were designed in such a way that the CPB and ECMO units were connected via the same circuitry. Such a connection allowed for swift switching between the 2 units when required throughout the course of entire procedure.

Introduction of the venous cannula to the right atrium (RA) through the right internal jugular vein was performed using the Seldinger technique. This cannula was used to drain blood from the RA to the ECMO circuit. With an incision parallel to inguinal ligament, the common femoral artery and vein were visualized and 5-0 purse-string closure sutures were placed in the common femoral artery and vein through which the arterial and venous cannula were introduced, respectively. A femoral drainage cannula should always be introduced into the RA (via the inferior vena cava) under transesophageal echocardiography supervision for safety precautions. The internal layer of each cannula is coated with heparin, which provides the ability to run the ECMO for more than 24 hours without the need for intravenous heparin.

Both venous (superior and inferior) cannulas were connected into 1 and then divided again into 2 lines with the use of a Y-shaped connector: 1 line for the ECMO circuit and 1 line for the CPB unit. An identical connection between the ECMO and CPB was performed with respect to the arterial cannula (Fig 4). Opening the

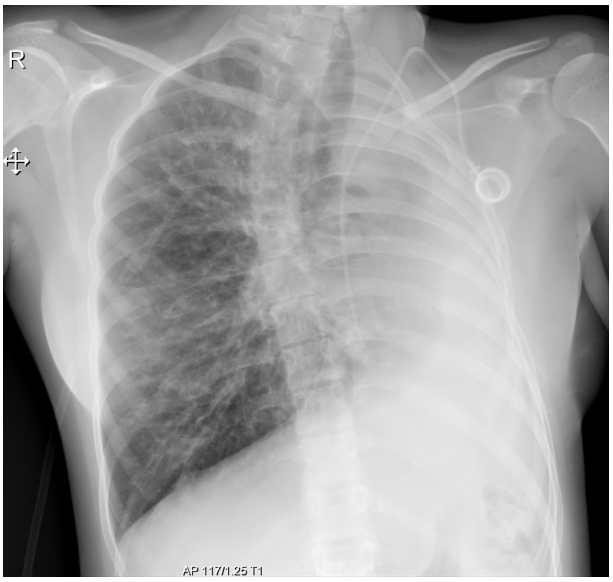

Fig 2. Chest radiograph before lung transplantation.

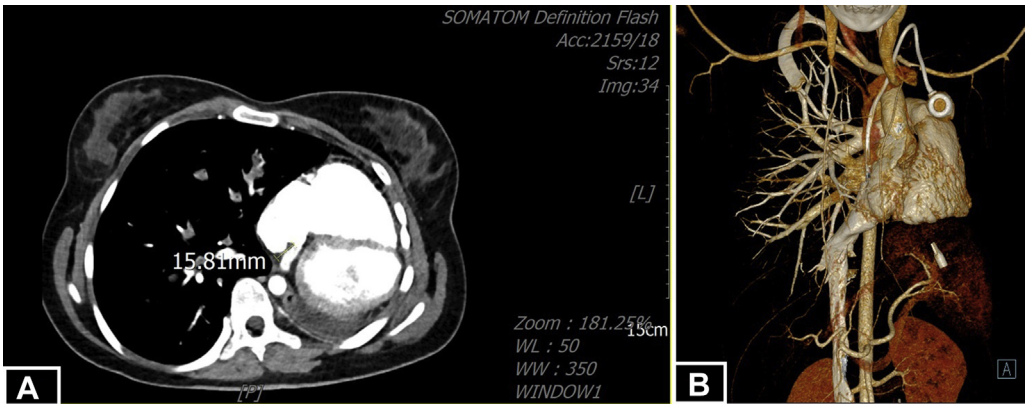

Fig 3. **(A)** Panel depicts the chest computed tomography with 1 measured thrombus. **(B)** Panel shows the 3-dimentional model of the patient's pulmonary circulation before the procedure. A vascular port device is present.

chest cavity (anterolateral thoracotomy in the fifth intercostal space on the right side) and preparation of the superior and inferior vena cava was the next step. The lung was dissected and cleaned of adhesions. The hilum of the lung was revealed and dissected. The pericardial sac was opened and holding sutures were placed on the superior and inferior vena cava. An adequate amount of heparin was administered to start CPB. The CPB unit and cardiotomy suction were turned on (with previous clamping of the ECMO lines [venous and arterial] past the Y-shaped connector). Holding sutures around the vena cava superior and vena cava inferior were tightened and the RA was opened.

Removal of the tumors from the RA was performed on a beating heart. When the RA was opened 4 mace-shaped tumors, each 1.0 to 3.0 cm in length, were revealed around ostium of the vena cava inferior and coronary sinus. They were excised and sent for histopathological examination. The RA was closed with 2 layers of 4-0 Prolene (Ethicon Inc, Somerville, NJ, United States) suture.

The next step was switching from CPB to ECMO. CPB and cardiotomy suction were stopped.

Lines to the CPB were clamped past the Y-shaped connector and the ECMO lines were unclamped.

Three-quarters of the full dose of protamine sulfate was administered and 200 seconds activated clotting time was set. Excision of the patient's lung and implantation of the donor's lung were performed using the standard approach. The final stage consisted of the explantation of the ECMO circuit and the CPB unit.

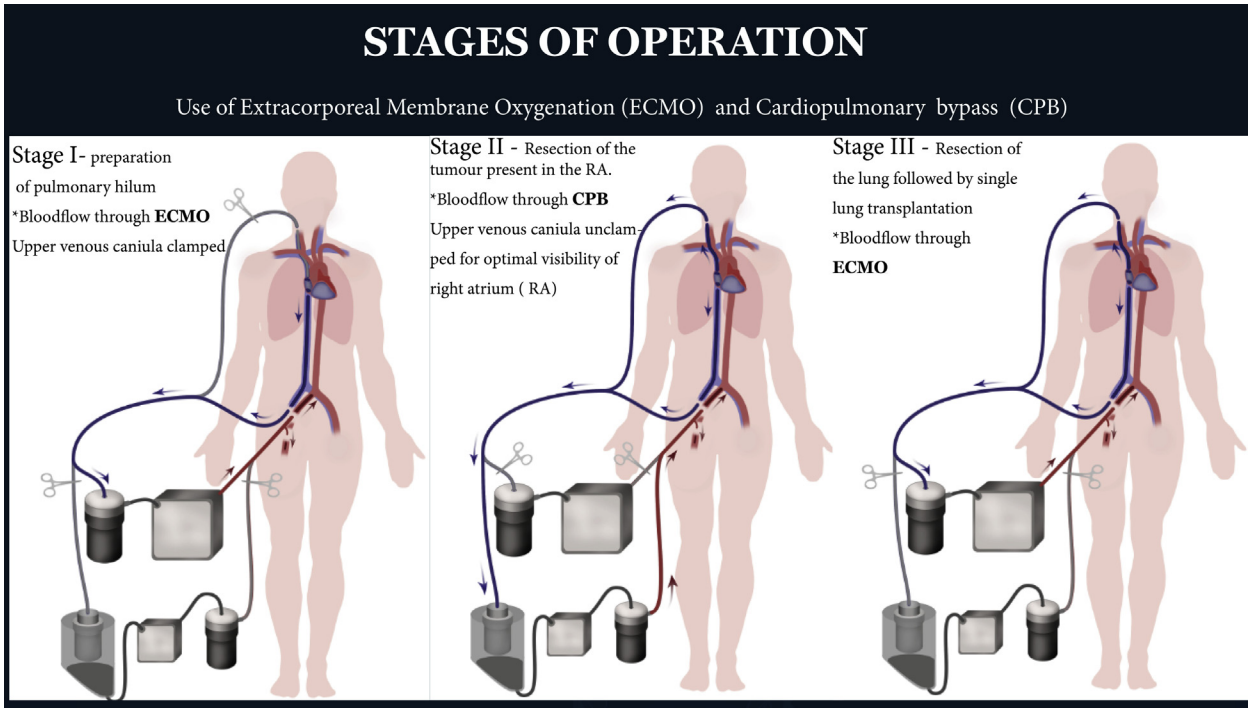

Fig 4. Stages of the operation. A full description is given in the text.

After closing the thoracotomy incision and finishing the main part of the procedure, both femoral cannulas (venous and arterial) were removed. Hemostasis and layered closure of the groin were performed. Removal of the venous cannula previously introduced through the jugular vein was uneventful [1 skin suture was placed using 4-0 Prolene (Ethicon Inc)]. The patient left the operating room without extracorporeal cardiopulmonary support.

Histologic examination of the removed tumors revealed that 3 of them were partially organized thrombi and the remaining fourth was a fibrotic thrombus.

Outcome after 1 Year

The patient remains under the care of our facility. She has already achieved 1-year survival with excellent spirometry results (47% of predicted forced expiratory volume in 1 second, 52% of predicted forced vital capacity, and proper Tiffeneau-Pinelli index of 77%). She also managed to walk 601 m during the 6-minute walk test with stable oxygen saturation of 97%. After the procedure, she gained weight and now presents with normal nutritional status. Her general condition improved and, as a result, she had her permanent venous access device explanted 6 months after the procedure. The rest of the posttransplant course is presented in Fig 1.

DISCUSSION

LT as a treatment for end-stage respiratory failure provides the best long-term survival among patients with CF according to data from the International Society for Heart and Lung Transplantation Registry [5]. Even though DLT seems to carry more benefits than SLT, this therapeutic option among some patients with CF is off limits. There was no technical possibility that our patient could have a DLT procedure because she underwent left-side pneumonectomy years prior to transplantation. Such treatment was reported to improve the patient's condition, as Liew et al reported in the case of a 14-year-old boy who underwent right pneumonectomy as a result of chronic history of collapse and consolidation of his entire right lung [6]. It was described that computed tomography revealed mild bronchiectasis of the left lung. After pneumonectomy, the patient regained good functional status. However, certain patients after pneumonectomy will not be eligible for DLT, as their pulmonary reserves will be diminished. The available literature reports cases of patients with CF who achieved satisfactory pulmonary capacity as recipients of SLT. Shennib et al reported a case of a recipient of DLT who underwent pneumonectomy 10 days after transplantation due to infarction of the left lung [7]. According to their case report, the patient is functioning extremely well. A standard DLT procedure allows the surgeon to explant the native lung first while ventilation is being maintained in the remaining lung on specific mechanical ventilation settings. In the case of patients who have had previous pneumonectomy, this procedure is not an option. Instead, oxygenation must be provided extracorporeally. A study published by Machuca et al found that intraoperative ECMO is a superior cardiopulmonary support [4]. However, our case required switching between these 2 devices, as the patient required ECMO during preparation of the pulmonary hilum followed by CPB support in order to perform removal of the masses from the RA. After successful completion of this part, blood flow was provided by the ECMO circuit in order to finish the LT. Cardiac operations concomitant with LT do not seem to increase a risk of death as reported by Shigemura et al, who noted several cases of patients who underwent tricuspid valvuloplasty followed by LT [8]. Their oxygenation and circulation support was provided by CPB during the valvuloplasty and implantation of the first lung. Switching from 1 method of cardiopulmonary support to another in the case of our patient was carried out carefully by an experienced team and provided an excellent outcome, as our patient is presently alive and well.

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